General

Guideline Title

Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management. A scientific statement from the American Heart Association.

Bibliographic Source(s)

Marino BS, Lipkin PH, Newburger JW, Peacock G, Gerdes M, Gaynor JW, Mussatto KA, Uzark K, Goldberg CS, Johnson WH Jr, Li J, Smith SE, Bellinger DC, Mahle WT, on behalf of the American Heart Association Congenital Heart Defects Committee [trunc]. Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association. Circulation. 2012 Aug 28;126(9):1143-72. [253 references] PubMed

Guideline Status

This is the current release of the guideline.

Recommendations

Major Recommendations

Definitions for the weight of the evidence (A-C) and classes of recommendations (I-III) are provided at the end of the "Major Recommendations" field.

Note from the American Heart Association (AHA) regarding language: For consistency, this statement uses terminology in accord with the 2006 American Academy of Pediatrics (AAP) policy statement on developmental surveillance and screening policy for the general pediatric population. Developmental "disorder" and "disability" (DD) are used equivalently within the context of this document and refer to the existence of a neurocognitive or neurobehavioral limitation or abnormality, psychosocial maladjustment, or physical limitation. In contrast, "development delay" is used to denote that a child's developmental maturation or "mental and/or physical skills are not consistent with the typical time frame." Surveillance, screening, and evaluation have distinct meanings and are defined as follows: (1) *Surveillance*—"the process of recognizing children who may be at risk for developmental delay"; (2) *screening*—"the use of standardized tools to identify and refine the risk" recognized from surveillance; and (3) *evaluation*—"a complex process aimed at identifying specific developmental disorders or disabilities that are affecting a child." The term *medical home* is per the 2002, 2005, and 2006 AAP policy statements and is "the optimal setting for family centered care coordination."

1. The medical home model of care may be effective and beneficial in the management of patients with chronic conditions such as congenital heart disease (CHD) (Class IIa; Level of Evidence B).
2. Existing AAP guidelines for surveillance, screening, evaluation, and intervention should be adhered to, with the following additions for patients with CHD:
    a. The following groups should be considered at high risk for DD (Class I; Level of Evidence A):
        1. Neonates or infants requiring open heart surgery (cyanotic and acyanotic types)
        2. Children with other cyanotic heart lesions not requiring open heart surgery in the neonatal or infant period
        3. Children with any combination of CHD and other comorbidities (see Table 3 of original guideline document)
        4. Other conditions determined at the discretion of the medical home providers
    b. Risk stratification of patients with CHD into low and high-risk categories for DD at every medical home visit can be useful and beneficial (Class IIa; Level of Evidence C).
    c. Behavioral screening of patients with CHD undergoing developmental screening based on age (9, 18, 30, 48 months) or concerns detected in surveillance (early childhood through adolescence) can be useful and beneficial (Class IIa; Level of Evidence C).

3. For patients with CHD stratified as being at high risk for DD, the following strategies can be useful and beneficial:
    a. Referral to formal developmental and medical evaluation can be useful and beneficial (Class IIa; Level of Evidence C).
    b. Referral to early intervention services or early childhood special education services before confirmation of a specific developmental diagnosis can be useful and beneficial (Class IIa; Level of Evidence B).
    c. Periodic reevaluations for DDs and developmental delays at 12 to 24 months, 3 to 5 years, and 11 to 12 years of age can be useful and beneficial (Class IIa; Level of Evidence C).
    d. Referral of young adults for higher education and/or vocational counseling can be useful and beneficial (Class IIa; Level of Evidence C).

Definitions:

Applying Classification of Recommendations and Level of Evidence

		SIZE OF TREATMENT EFFECT					
		CLASS I *Benefit >>> Risk* Procedure/Treatment SHOULD be performed/ administered	CLASS IIa *Benefit >> Risk Additional studies with focused objectives needed* IT IS REASONABLE to perform procedure/administer treatment	CLASS IIb *Benefit ≥ Risk Additional studies with broad objectives needed; additional registry data would be helpful* Procedure/Treatment MAY BE CONSIDERED	CLASS III *No Benefit* or CLASS III *Harm*		
						Procedure/Test	Treatment
					COR III: No benefit	Not Helpful	No Proven Benefit
					COR III: Harm	Excess Cost w/o Benefit or Harmful	Harmful to Patients
Estimate of Certainty (Precision) of Treatment Effect	LEVEL A Multiple populations evaluated* Data derived from multiple randomized clinical trials or meta-analyses	• Recommendation that procedure or treatment is useful/effective • Sufficient evidence from multiple randomized trials or meta-analyses	• Recommendation in favor of treatment or procedure being useful/effective • Some conflicting evidence from multiple randomized trials or meta-analyses	• Recommendation's usefulness/efficacy less well established • Greater conflicting evidence from multiple randomized trials or meta-analyses	• Recommendation that procedure or treatment is not useful/effective and may be harmful • Sufficient evidence from multiple randomized trials or meta-analyses		
	LEVEL B Limited populations evaluated* Data derived from a single randomized trial or nonrandomized studies	• Recommendation that procedure or treatment is useful/effective • Limited evidence from single randomized trial or nonrandomized studies	• Recommendation in favor of treatment or procedure being useful/effective • Some conflicting evidence from single randomized trial or nonrandomized studies	• Recommendation's usefulness/efficacy less well established • Greater conflicting evidence from single randomized trial or nonrandomized studies	• Recommendation that procedure or treatment is not useful/effective and may be harmful • Evidence from single randomized trial or nonrandomized studies		
	LEVEL C Very limited populations evaluated* Only consensus opinion of experts, case studies, or standard of care	• Recommendation that procedure or treatment is useful/effective • Only expert opinion, case studies, or standard of care	• Recommendation in favor of treatment or procedure being useful/effective • Only diverging expert opinion, case studies, or standard of care	• Recommendation's usefulness/efficacy less well established • Only diverging expert opinion, case studies, or standard of care	• Recommendation that procedure or treatment is not useful/effective and may be harmful • Only expert opinion, case studies, or standard of care		

A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Although randomized trials are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective (see Table 1 in the original guideline document for a list of suggested phrases for writing recommendations).

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as sex, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use.

Clinical Algorithm(s)

The original guideline document provides the following clinical algorithm: "Congenital heart disease (CHD) algorithm for surveillance, screening, evaluation, and management of developmental disorders and disabilities" (see Figure A and B).

Note: The algorithm complements the general algorithm from the American Academy of Pediatrics 2006 policy statement entitled, "Identifying Infants and Young Children with Developmental Disorders in the Medical Home: An Algorithm for Developmental Surveillance and Screening."

Scope

Disease/Condition(s)

- Developmental disorders or disabilities
- Developmental delay
- Congenital heart disease (CHD)

Guideline Category

Evaluation

Management

Rehabilitation

Risk Assessment

Screening

Clinical Specialty

Cardiology

Family Practice

Medical Genetics

Neurology

Nursing

Pediatrics

Physical Medicine and Rehabilitation

Psychiatry

Psychology

Speech-Language Pathology

Intended Users

Advanced Practice Nurses

Health Care Providers

Nurses

Occupational Therapists

Physical Therapists

Physicians

Psychologists/Non-physician Behavioral Health Clinicians

Speech-Language Pathologists

Guideline Objective(s)

- To review the factors underlying the increased risk for developmental delays and disabilities (DD) in the congestive heart disease (CHD) population, recommend a CHD algorithm for DD that incorporates risk stratification, review age-based management of CHD patients, and discuss the impact of DD on quality of life (QOL) for the CHD population
- To provide a new framework for the surveillance, screening, evaluation, and management of DDs in the pediatric CHD population

Target Population

Children and adolescents with congenital heart disease (CHD) in the medical home setting

Interventions and Practices Considered

1. Use of the medical home model of care
2. Surveillance, screening, evaluation, and intervention strategies in accordance with American Academy of Pediatrics policies
3. Determination of infants and children at high risk for development delays and disabilities (DD)
4. Risk stratification of patients with congenital heart disease (CHD) into low and high-risk categories for DD at every medical home visit
5. Behavioral screening of patients with CHD undergoing developmental screening based on age or concerns detected in surveillance
6. Referral of high-risk patients to formal developmental and medical evaluation, early intervention services, or early childhood special education services
7. Periodic reevaluations for DDs and developmental delays
8. Referral of young adults for higher education and/or vocational counseling

Major Outcomes Considered

- Risk of developmental delays and disabilities (DDs), academic difficulties, behavioral abnormalities, and psychosocial problems in infants and children with congenital heart disease (CHD)
- Effectiveness of screening approaches, including behavioral and psychosocial, for patients with CHD
- Effectiveness of referral to interventions to improve diagnosis and/or developmental outcomes in patients with CHD
- Impact of DD on quality of life

Methodology

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Hand-searches of Published Literature (Secondary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

MEDLINE and Google Scholar database searches from 1966 to 2011 were conducted for English-language articles cross-referencing congenital heart disease (CHD) with pertinent search terms (i.e., attention deficit hyperactivity disorder, autism spectrum disorders, brain injury, behavioral issues, cardiopulmonary resuscitation, developmental disorder, developmental disability, developmental delay, developmental screening, fine and gross motor abnormalities, genetic disorder or syndrome, heart transplantation, mechanical support, microcephaly, neurodevelopment, neurodevelopmental outcome, periventricular leukomalacia, prematurity, prolonged hospitalization, psychological issues, psychosocial abnormalities, quality of life, seizures, stroke, transition, and adult CHD). The reference lists of identified articles were also searched. Published abstracts from major pediatric scientific meetings in 2010 and 2011 were also reviewed.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Applying Classification of Recommendations and Level of Evidence

		SIZE OF TREATMENT EFFECT					
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Estimate of Certainty (Precision) of Treatment Effect	LEVEL A Multiple populations evaluated* Data derived from multiple randomized clinical trials or meta-analyses	• Recommendation that procedure or treatment is useful/effective • Sufficient evidence from multiple randomized trials or meta-analyses	• Recommendation in favor of treatment or procedure being useful/effective • Some conflicting evidence from multiple randomized trials or meta-analyses	• Recommendation's usefulness/efficacy less well established • Greater conflicting evidence from multiple randomized trials or meta-analyses	• Recommendation that procedure or treatment is not useful/effective and may be harmful • Sufficient evidence from multiple randomized trials or meta-analyses		
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A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical

trials. Although randomized trials are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective (see Table 1 in the original guideline document for a list of suggested phrases for writing recommendations).

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as sex, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use.

Methods Used to Analyze the Evidence

Review of Published Meta-Analyses

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

Classification of recommendations and level of evidence were assigned to each recommendation per the manual for American College of Cardiology (ACC)/American Heart Association (AHA) guideline writing committees ("Methodologies and Policies From the ACC/AHA Task Force on Practice Guidelines," section 4: writing recommendations; see the "Availability of Companion Documents" field). The ACC/AHA guidelines grading schema based on level of evidence and class of recommendation (see "Rating Scheme for the Strength of the Evidence") were used. The level of evidence classification combines an objective description of the existence and the types of studies that support the recommendation and expert consensus.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

A writing group appointed by the American Heart Association and American Academy of Pediatrics reviewed the available literature addressing developmental disorder and disability and developmental delay in the congenital heart disease (CHD) population, with specific attention given to surveillance, screening, evaluation, and management strategies. A CHD algorithm for surveillance, screening, evaluation, reevaluation, and management of developmental disorder or disability has been constructed to serve as a supplement to the 2006 American Academy of Pediatrics statement on developmental surveillance and screening.

Rating Scheme for the Strength of the Recommendations

See the "Rating Scheme for the Strength of the Evidence" field, above.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

Expert peer review of American Heart Association (AHA) Scientific Statements is conducted by the AHA Office of Science Operations.

The statement was approved by the American Heart Association Science Advisory and Coordinating Committee on April 27, 2012.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

- Incorporation of a new stratification method and clinical algorithm may result in increased surveillance, screening, evaluation, diagnosis, and management of developmental disorders or disabilities (DDs) in the complex congenital heart disease (CHD) population and consequent improvement in neurodevelopmental and behavioral outcomes in this high-risk population. With early identification of DDs and developmental delays, children have the best chance to reach their full potential.
- Periodic developmental surveillance, screening, evaluation, and reevaluation throughout childhood may enhance identification of significant deficits, allowing for appropriate therapies and education to enhance later academic, behavioral, psychosocial, and adaptive functioning.

Potential Harms

Not stated

Qualifying Statements

Qualifying Statements

This statement has not been formally disseminated by the Centers for Disease Control and Prevention/the Agency for Toxic Substances and Disease Registry. It does not represent and should not be construed to represent any agency determination or policy.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Clinical Algorithm

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Living with Illness

Staying Healthy

IOM Domain

Effectiveness

Patient-centeredness

Timeliness

Identifying Information and Availability

Bibliographic Source(s)

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Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2012 Aug 28

Guideline Developer(s)

American Heart Association - Professional Association

Source(s) of Funding

American Heart Association

Guideline Committee

American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Stroke Council

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Financial Disclosures/Conflicts of Interest

The American Heart Association makes every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest.

Writing group and reviewer disclosures can be found at the end of the original guideline document.

Guideline Endorser(s)

American Academy of Pediatrics - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the American Heart Association Web site .

Print copies: Available from the American Heart Association, Public Information, 7272 Greenville Ave, Dallas, TX 75231-4596; Phone: 800-242-8721

Availability of Companion Documents

The following is available:

- Methodology manual and policies from the ACCF/AHA Task Force on Practice Guidelines. 2010 Jun. 88 p. American College of Cardiology Foundation (ACCF) and American Heart Association (AHA), Inc. Electronic copies: Available in PDF from the AHA Web site .

Patient Resources

None available

NGC Status

This summary was completed by ECRI Institute on September 28, 2012. The information was verified by the guideline developer on October 22, 2012.

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